**A Rare Complication of a Rare Disease: Pericarditis Due to Ruptured Pericardial Cyst**

Hassan Lak, MDª, Rohan Shah, MDᵇ, Beni Rai Verma, MDª, Bo Xu, MDb, c

ª Department of Medicine, Cleveland Clinic, Cleveland, Ohio, USA, 44195

ᵇ Robert and Suzanne Tomsich Department of Cardiovascular Medicine, Heart, Vascular and Thoracic Institute, Cleveland Clinic, Cleveland, Ohio, USA, 44195

ͨ Section of Cardiovascular Imaging, Robert and Suzanne Tomsich Department of Cardiovascular Medicine, Heart, Vascular and Thoracic Institute, Cleveland Clinic, Cleveland, Ohio, USA, 44195

**Address for correspondence**

Bo Xu, MD, FRACP, FACC, FASE

Department of Cardiovascular Medicine, Heart, Vascular and Thoracic Institute, Cleveland Clinic

9500 Euclid Avenue, Desk J1-5, Cleveland, Ohio, 44195

Phone: 216.444.2200

Fax: 215.445.6152

Email: xub@ccf.org

Funding: No funding sources

Disclosures: The authors have no disclosures

**Abstract**

Pericardial cysts are considered rare incidental findings, which are generally asymptomatic in nature. Occasionally, patients may represent with chest discomfort, dyspnea or palpitations. Pericarditis related to a ruptured pericardial cyst has not been previously reported in the literature. Here, we report the case of a 62-year-old male who developed acute pericarditis as a result of a ruptured enlarging pericardial cyst.

**Keywords**: pericarditis; pericardial cyst; echocardiography; computed tomography; cardiac magnetic resonance imaging

Word count: 1577

Figures: 3

**Abbreviations**:

CMR: Cardiac Magnetic Resonance

CTPA: Computed Tomography Pulmonary Angiogram

NT-Pro BNP: NT-Pro Brain-derived Natriuretic Peptide

ESR: Erythrocyte Sedimentation Rate

CRP: C-reactive Protein

HFrEF: Heart Failure with Reduced Ejection Fraction

**Introduction**

Pericardial cysts are rare mediastinal masses with a reported incidence of 1 in 100,000 patients.1 They are mostly asymptomatic and detected incidentally, but can present with chest discomfort, dyspnea or palpitations. They are often found in one of the cardiophrenic angles and can be uni- or multiloculated.1 These cysts are usually classified into two categories: congenital and acquired.2 We report a case of 62 year-old male with a history of large right-sided pericardial cyst who presented with chest discomfort, new-onset of shortness of breath and bilateral leg swelling. Recent chest computed tomography (CT) performed to exclude pulmonary embolism prior to the current presentation, demonstrated a right-sided pericardial cyst measuring 8.7 x 2.5 cm which was enlarging, in comparison to a prior CT (dimensions 7.3 x 1.7 cm). Upon admission, transthoracic echocardiography performed showed a left ventricular ejection fraction of 25% and dilated cardiac chambers. Elective coronary angiogram showed no significant coronary artery disease. Cardiac magnetic resonance (CMR) revealed that the previously present pericardial cyst was no longer present. However, severe delayed pericardial enhancement consistent with active inflammation was evident. The patient’s condition improved after initiation of anti-inflammatory therapy with high dose aspirin and colchicine. This case showed that the rupture of a pericardial cyst may result in acute pericarditis. Multimodality imaging with computed tomography and cardiac magnetic resonance play an essential role in timely diagnosis and initiation of treatment.

**Case Presentation**

A 62-year-old male with a known history of a right-sided pericardial cyst was admitted to our hospital with chest discomfort, New York Heart Association (NYHA) class III dyspnea and leg swelling of two months. He also experienced fatigue and diaphoresis. On examination, he was afebrile, blood pressure was 98/57 mmHg, pulse rate was 87 beats per minute, respiratory rate was 16 per minute and oxygen saturation was 94% on room air. On chest auscultation, S1 and S2 were heard, with no murmurs or pericardial rub. Breath sounds were normal without crackles. Patient had a past medical history of a known right sided pericardial cyst and alcohol abuse. Differential diagnosis included acute systolic or diastolic heart failure, acute myocardial infarction or pericarditis.

**Investigations**

His initial laboratory investigations demonstrated a normal troponin I to 0.028 ng/ml (reference range 0.000-0.029 ng/ml) and elevated NT-Pro brain-derived natriuretic peptide (NT-Pro BNP) at 2840 pg/ml (reference range < 125 pg/ml). 12-lead electrocardiogram showed normal sinus rhythm with no ischemic changes. White blood cell count was 14.11 k/ul (reference range: 3.70-11.00 k/ul), erythrocyte sedimentation rate (ESR) of 35 mm/hour (reference range: 0-20 mm/hr) and ultrasensitive C-reactive protein (CRP) of 48.4 mg/dl (reference range <3.1 mg/L). Transthoracic echocardiography demonstrated a left ventricular ejection fraction of ~25% and no significant valvular dysfunction. Computed tomography (CT) pulmonary angiogram performed 1 month prior to admission, demonstrated that his right-sided pericardial cyst, measured 8.7 x 2.5 cm (compared to 7.3 x 1.7 cm at time of initial diagnosis of the pericardial cyst, 4 years prior) (Figure 1).

**Management**

The patient was admitted to the cardiology unit, and initial medical regimen included carvedilol, spironolactone, losartan and furosemide. In order to evaluate for systolic heart failure, coronary angiography was performed which showed no significant atherosclerotic disease. Further investigation with cardiac magnetic resonance imaging (CMR) – delayed enhancement imaging revealed no evidence of ischemic myocardial damage or interstitial fibrosis, and surprisingly, the previously known pericardial cyst was no longer existent. There was markedly increased pericardial signal, consistent with active pericardial inflammation (Figure 2a). Black blood imaging demonstrated markedly thickened pericardium (Figure 2b) and T2-weighted short-tau inversion recovery (T2-STIR) edema-weighted imaging showed significant pericardial edema (Figure 3). There was no significant diastolic septal bounce, diastolic restraint, or conical deformity of the ventricles to suggest constrictive physiology. The patient was started on high dose aspirin (650 mg tablets twice daily) and colchicine (0.6 mg tablets twice daily) and discharged home.

**Outcome and Follow-up**

At 2-month follow-up, his dyspnea improved to NYHA functional class I. Repeat ultra-sensitive CRP was 4.0 mg/dl (reference range <3.1 mg/L). The patient is alive and well at 6 month follow-up.

**Discussion**

Anatomically, pericardium is formed of a visceral and a parietal layer surrounding the heart. It receives blood supply from phrenic arteries, lymphatic drainage into mediastinal lymph nodes and innervation by the phrenic nerve.3,4 Lymphatics from anterior and posterior parietal pericardium drain into anatomically corresponding mediastinal nodes.3 The superficial plexus of cardiac lymphatics provides drainage for the visceral pericardium draining into tracheal and bronchomediastinal lymph nodes.3 Between the visceral and parietal pericardium is an echolucent space containing 15 and 50 mL of serous fluid, which in the normal state represents physiologic pericardial effusion (Peff).5

Pericardial cysts are rare mediastinal masses with a reported incidence of 1 in 100,000 patients.1 These are detected incidentally during evaluation for other clinical conditions. They typically occur along the right heart border (70% cases), but can also occur at other locations. They are generally asymptomatic, causing symptoms only if adjacent structures (e.g., coronary arteries) are compressed.1When symptomatic, patients may develop shortness of breath due to right sided heart failure secondary to compression.7,8 Clinical significance of pericardial cysts lies in the fact that they need to be distinguished from other cardiac and mediastinal masses.

Pericardial cysts can be classified into two categories: congenital and acquired.9 Embryologically, the pericardium arises from lacunae that gradually merge and form the pericardial cavity. If they fail to unite fully, a pericardial cyst may develop.2 Acquired pericardial cysts can occur due to infections, with the leading cause being a rare complication of Echinococcosis hydatid cysts.2 Other acquired causes include malignant metastasis, post-traumatic, pericarditis, cardiac surgery and rheumatic heart disease.10 There is limited literature showing that ruptured pericardial cyst may present with acute chest pain syndrome.10 To the best of our knowledge, pericarditis complicating a ruptured pericardial cyst has not been previously reported in the literature. The presence of a pericardial cyst is usually suggested by the chest radiograph. Pericardial cysts, may be difficult to detect with transthoracic echocardiography and can appear as an echo-free space, localized and spherical area. On cardiac CT imaging, pericardial cysts appear as non-enhancing, extracardiac, intrapericardial, homogenous cystic lesions.8 On CMR imaging, pericardial cysts are identified as homogenous, hyperintense lesions on T1 and T2-weighted sequences.8 Either cardiac CT or CMR can be used to confirm its presence in case of clinical suspicion.

In our case, CMR imaging of the pericardium showed inflammatory findings consistent with active pericarditis. It also suggested rupture of the previously documented pericardial cyst, which was previously documented to have progressively increased in size. It appears from clinical presentation that the patient developed rupture of the enlarging pericardial cyst which was further complicated by acute pericarditis. Hence, this case highlights a previously unreported complication of a rare entity.

**Conclusion**

Pericardial cysts are rare mediastinal cysts which can be congenital or acquired. In this case, the rupture of a pericardial cyst has resulted in acute pericarditis. This was successfully treated with oral anti-inflammatory therapy. Multimodality imaging with computed tomography and cardiac magnetic resonance is useful in diagnosing and managing complications related to pericardial cysts. Though generally rare and thought to be asymptomatic, this case highlights that when patients with pericardial cysts present with new symptoms, careful evaluation and imaging-guided evaluation is warranted to exclude complications.

**Learning Objectives**

• Pericardial cyst are rare mediastinal masses which are most commonly detected incidentally

• They are mostly asymptomatic, but have been reported to cause symptoms secondary to compression of surrounding structures

• It is important to differentiate them from other mediastinal masses

• Acute pericarditis is an extremely rare complication of a ruptured pericardial cyst

**Figures**

**Figure 1**: Chest computed tomography (CT) (1a) at the time of initial discovery of the pericardial cyst, measuring 7.3 x 1.7 cm; (1b) repeat computed tomographic imaging 4 years later showed the pericardial cyst was enlarging, now measuring 8.7 x 2.5 cm.

**Figure 2**: CMR phase sensitive inversion recovery (PSIR) sequence showing severe delayed enhancement of the pericardium (arrows), consistent with active pericardial inflammation (2a). Black blood axial imaging, demonstrating markedly thickened pericardium measuring 5 mm (2b).

**Figure 3**: Cardiac magnetic resonance (CMR) with T2-weighted short tau inversion recovery (STIR) sequence, showing significant pericardial edema, consistent with active pericarditis.

**References**

1. Gouriet F, Levy P, Casalta J et al. Etiology of Pericarditis in a Prospective Cohort of 1162 Cases. *Am J Med*. 2015;128(7):784.e1-784.e8. doi:10.1016/j.amjmed.2015.01.040

2. Khayata M, Alkharabsheh S, Shah NP, Klein AL. Pericaridal cysts: a contemporary comprehensive review. *Curr Cardiol Rep*. 2019;21(7):64. doi: 10.1007/s11886-019-1153-5

3. Spodick D. The normal and diseased pericardium: Current concepts of pericardial physiology, diagnosis and treatment. *J Am Coll Cardiol*. 1983;1(1):240-251. doi:10.1016/s0735-1097(83)80025-4

4. Xu B, Kwon DH, Klein AL. Imaging of the Pericardium: A Multimodality Cardiovascular Imaging Update. *Cardiol Clin*. 2017;35(4):491-503. doi: 10.1016/j.ccl.2017.07.003

5. Spodick D. Macrophysiology, microphysiology, and anatomy of the pericardium: A synopsis. *Am Heart J*. 1992;124(4):1046-1051. doi:10.1016/0002-8703(92)90990-d

6. Kittredge RD, Finby N. Pericardial cysts and diverticula. *American Journal of Roentgenology*. 1967;99(3):668-673. doi:10.2214/ajr.99.3.668

7. Valschaerts A, Dupont M, Seldrum S. Idiopathic pericardial cyst causing heart failure. *Acta Cardiol*. 2018;74(2):174-175. doi:10.1080/00015385.2018.1453450

8. Bastarrika G, De Cecco C, Arraiza M et al. Morphological and functional evaluation of intrapericardial cyst as a cause of severe right heart failure: dual source computed tomography and magnetic resonance imaging. *Journal of Cardiovascular Medicine*. 2009;10(4):363-364. doi:10.2459/jcm.0b013e32832607ca

9. Roberts W. Pericardial Heart Disease: Its Morphologic Features and Its Causes. *Baylor University Medical Center Proceedings*. 2005;18(1):38-55. doi:10.1080/08998280.2005.11928030

10. Aertker R, Cheong B, Lufschanowski R. Inflammation and Rupture of a Congenital Pericardial Cyst Manifesting Itself as an Acute Chest Pain Syndrome. *Tex Heart Inst J*. 2016;43(6):537-540. doi:10.14503/thij-15-5623